1072-98 Two Thousand Adults With Congenital Heart Disease: Operative and Nonoperative Mortality

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Background: All 2000 adults with congenital heart disease (CHD) seen between 1987 - 2000 were evaluated for all-cause mortality.
Methods: Prospective data acquisition of all patients (pts) seen in the Adult CHD Clinic at a single center.
Results: Of 2000 pts, age 13 - 85 yrs (mean 37.1), 752 (38%) were ≥ 40 yrs old at referral; 951 had never had surgery. One thousand and fifty-four operations were performed between 1987-2000. A total of 167 deaths (8%) occurred during follow-up (age 17- 89 yrs, mean 42.9) Thirty-nine deaths (23%) were peri-operative (< 30-day: p-op); 3 p-op deaths occurred elsewhere. Eleven of our 36 p-op deaths (31%) occurred at initial operation ;8 Ebsteins (7 with advanced CHD at referral and 1 unexpected), and 3 complex pulmonary atresia (PA) (2 with advanced CHD and 1 sub-optimal selection). Of 25 deaths with re-operation (re-op), 12 had advanced CHD and were high-risk (3 with PA, 2 with aortic stenosis, 2 with single ventricle physiology and 1 each Ebstein, tetralogy, truncus, complex anomalous venous return and pulmonary hypertensive ductus/coarctation). Nine re-op deaths were unexpected (3 strokes, 2 bleeds, 2 cardiac failure (CHF), 1 rejection after transplant and 1 spontaneous coronary dissection), 2 had sub-optimal selection (1 PA, 1 Fontan) and 2 had endocarditis. Seventeen p-op deaths occurred in pts having operation number ≥ 3 (range 3-8). Of 128 non-surgical deaths, 82 were expected cardiovascular (CV) : 34 with advanced CHD, 27 with CHF and 21 with Eisenmenger syndrome (7 sudden). Twenty-three pts died suddenly (7 with tetralogy).Four of these pts.(2 with d-transposition and 2 with tetralogy) had modest conduit obstruction. Nine pts died from cancer, 3 pts died after non-CV surgery (2 Eisenmenger, 1 Fontan) and 10 from miscellaneous causes.
Conclusions: In this older cohort, a low op mortality is possible even in pts having multiple re-ops. Most p-op deaths (21/36, 58%) relate to advanced CHD and late referral. Early recognition and referral to tertiary centers is recommended. Most non-surgical deaths also relate to advanced disease (82/128, 64%) although risk stratification for sudden death remains a challenge.

1072-99 How Important Is Cardiac MRI in the Management of Adults With Congenital Heart Disease?

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Background: Assessment of RV function and complex anatomy with echocardiography in adults with congenital heart disease (CHD) can be difficult and may require other imaging modalities. Cardiac MRI is frequently used to answer these questions. We sought to determine the value of cardiac MRI in patients with insufficient echocardiographic information to determine its effect on subsequent management. Methods: We retrospectively reviewed all adults with CHD who underwent both echocardiography (Echo) and MRI at New York Presbyterian Hospital from 7/99 to 8/01. Indications for study were: (1) assessment of RV function (Group I) and (2) delineation of anatomy (Group II). Studies were performed on a 1.5T magnet utilizing cardiac gated spin echo and multiplanar cine with or without breath hold. Contrast enhanced MRA was utilized selectively to visualize arterial or venous abnormalities. Results: There were 66 pts (42M 24F) with a mean age of 32 years (range 18-61). There were 28 pts in Group I; 23 with Tetralogy of Fallot (TOF) and pulmonary regurgitation and 5 with D or L-transposition of the great arteries. MRI was discordant with echo in 5 of 23 TOF pts (22%), all of whom had better RV function on MRI which delayed pulmonary valve replacement (PVR). In 6 additional pts (21%) in Group I, MRI revealed unsuspected anatomy that led to catheter intervention in 4. There were 38 pts in Group II, 12 of whom (32%) had MRI findings which led to catheter intervention, surgery or identification of a potential surgical problem. In 8 pts with prior coarctation repairs, MRI delineated the site for stent insertion in 5 and revealed an unsuspected false aneurysm in 2 pts requiring surgery. Overall, MRI resulted in catheter intervention or surgery in 14 of 66 pts (21%) and delayed PVR in 22% of TOF pts with pulmonary regurgitation. MRI failed to identify a venous anomaly in 2 pts referred for assessment of RV function in which MRA was not performed. Conclusions: In adults with CHD, cardiac MRI provides structural and functional information not obtainable with transthoracic echocardiography that can lead to intervention or affect the decision for PVR. Use of MRI in selected lesions can facilitate the management of adults with CHD.

1072-100 Beneficial Effects of Atrial Septal Defect Percutaneous Closure in Asymptomatic and Mildly Symptomatic Adults

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Background: Controversy exists as to whether secundum type atrial septal defects (ASD) in asymptomatic or mildly symptomatic (NYHA I-II) adult patients (pts) should be closed.
Methods: Twenty-two pts (15 females; mean age 48.5 years, 19 to 74) with a Qp/Qs of 2.2/1 (1.4 to 3.4/1) had a VO2max determination and transthoracic echocardiogram before and 6 months after elective closure of ASD with an Amplatzer device. Short (SAX) and long (LAX) axis dimensions of the right ventricle were measured in the 4-chamber view. Right ventricular myocardial performance index (MPI) was calculated.
Results: At baseline, mean VO2max was 25.1 cc/kg/min and was higher in NYHA I pts than in NYHA II pts (29.2 vs 21.9 cc/kg/min; p<0.02). Qp/Qs was higher in NYHA II pts; (2.4/1 vs 1.8/1; p<0.05) but there was no difference in right ventricular dimensions. VO2max increased significantly at 6 months (25.1 to 28.4 cc/kg/min; p=0.0002). Improvement was as marked in the 9 NYHA I pts (+13.7%; from 29.3 to 33.1) as in the 13 NYHA II pts (+12.3%; 21.9 to 24.6), in pts with Qp/Qs 1.4 to 2.0 (+11.7%) as in those with Qp/Qs greater or equal 2.0 (+12.4%) and in pts greater or equal 50 years (+11.7%) as in those <50 years (+16.2%). Compared to 9/22 pts preprocedure, 21/22 pts were in NYHA I at 6 months. SAX decreased from 45 to 35 mm (p<0.0001) and LAX from 75 to 66 mm (p=0.0005). MPI was normal at baseline and at 6 months (0.20 to 0.36).
Conclusion: 1) Adult ASD pts significantly increase their maximal O2 consumption after percutaneous defect closure.
2) This can be observed even in pts classified as asymptomatic, in those with lesser shunts and in older pts.
These findings suggest that ASD closure in an adult population with a Qp/Qs >1.4 should be considered even in the absence of symptoms.

1072-101 Incidence of Infective Endarteritis in Adults With a Patent Ductus Arteriosus

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Background
The contemporary risk of infective endarteritis (IE) in adult patients with a patent ductus arteriosus (PDA) is unclear. The aim of this study was to estimate the incidence of IE in adult patients with PDA and ascertain associated risk factors.
Methods
Patients with a diagnosis of PDA, not associated with complex congenital heart disease, were identified from the University of Toronto Congenital Cardiac Centre for Adults Cardiology database. Clinical records were reviewed to determine the prevalence of symptoms, and arrhythmia, the presence of a typical murmur, and echocardiographic indices of left ventricular function. PDA size and morphology were determined angiographically in patients considered for device closure. Hospital records were reviewed to identify cases of possible or definite IE. Patients were considered at risk from age 16 until either closure of the PDA or latest clinical review. Logistic regression analysis was used to determine risk factors for IE.
Results
175 cases of PDA were identified (median age 37 [range 16-83] years, 79% female). A murmur was present in 70% although 57% were asymptomatic. Symptoms included dyspnoea (40%) and palpitations (12%). Clinical heart failure and arrhythmia were present in 8.2%. and 5.8% respectively. Five cases of definite IE were reported during a total of 6897 years of follow-up though only 4 of these occurred during 4097 years of adult life (incidence 0.1%/year>16yrs old). All patients with IE had a murmur though one was asymptomatic with no evidence of left heart volume load on echocardiography. Duct sizes in patients with IE were 3.7, 4, 5.5, and 7.2 mm. No significant risk factors for IE were identified from baseline data.
Conclusion
These data indicate a low risk of IE in adult patients with PDA. However, predicting those at risk from IE remains difficult and it may develop in patients with small PDAs and no clinical evidence of left heart volume load.

1072-102 Heart Transplantation in Adolescents and Adults With Congenital Heart Disease

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Background: There are limited data on outcomes following heart transplantation in adolescents and adults with congenital heart disease (CHD). Accordingly, we assessed potential relationships between pre and post-transplantation risk factors and morbidity/mortality.
Methods: Twenty nine patients with congenital heart disease over age 13 years underwent orthotopic heart transplantation since 1984 out of a total of 1148 patients. Thirteen were aged 13 to 17 years, 16 were aged 18 to 49 years, and the mean age at transplantation was 23 years. The most frequent pre-transplant CHD diagnosis was single ventricle (n= 14) followed by d-transposition of the great arteries (n = 9). Twenty-seven of 29 patients had 1 to 5 prior cardiac operations (mean 2). Of the 27, 13 had a Fontan procedure, 3 had a Mustard or Senning procedure, and 11 had palliative shunts. The primary indication for transplantation was refractory heart failure.
Results: Early deaths occurred in 6/ 29 (20.6%), and late deaths occurred in 4/23 (17.4%) during a mean follow up of 5.8 years. Four of 6 early deaths were due to surgical bleeding mainly from extensive dissection, one to acute rejection, and one to refractory ventricular tachyarrhythmias. Extensive reconstruction significantly prolonged the ischemic time. Three of 4 late deaths were among the 4 patients who developed lymphoproliferative disorders. The fourth late death was due to multi-organ failure following re-transplantation for transplant vasculopathy. More patients experienced recurrent episodes of acute rejection in the first three years in contrast to transplant patients without congenital heart disease.
Conclusions: Heart transplantation can be performed successfully in most adolescents and adults with congenital heart disease. However, an increased risk of bleeding is incurred because of extensive adhesions or collaterals. The higher rejection rate may be related to multiple blood transfusions during many pre-transplantation surgeries. Lymphoproliferative disorders may occur with increased frequency as a consequence of the combined effects of aggressive immunosuppresion and increased pre-transplantation radiation exposure.